

Surgical management of Grisel syndrome in the adult patient: illustrative case

*Mohamed Macki, MD, MPH, Jacob Pawloski, MD, Hassan A. Fadel, MD, and Muwaffak M. Abdulhak, MD

Department of Neurosurgery, Henry Ford Hospital, Detroit, Michigan

BACKGROUND Grisel syndrome describes an infectious soft tissue process that destabilizes the cervical bony elements and ligamentous complexes. This nontraumatic atlantoaxial rotary subluxation occurs in children primarily. This case illustrates a rare case presentation of an adult with Grisel syndrome: infectious destruction of the right atlantoaxial facet joint caused the occiput-C1 vertebra (head) to rotate rightward with lateral horizontal displacement off the C2 vertebra.

OBSERVATIONS Because the infection destroyed the C1 bony arch and atlantoaxial facet joints with epidural extension, the rotated head and atlas pulled the brainstem–cervical spinal cord junction against a fixed odontoid process, resulting in a cord contusion. Because of the highly unstable craniocervical junction, the patient presented with torticollis and left upper extremity weakness.

LESSONS Treatment entailed closed reduction under general anesthesia followed by occipitocervical fusion with an occipital plate, C1 lateral mass screws, and C2–C5 pedicle screws. This case describes the unique surgical pearls necessary for occipitocervical fusion of an unstable craniocervical junction, including tips with neuronavigation, trajectories of the cervical pedicle screws, aligning the lateral mass and pedicle screws with the occipital plate, and nuances with occipitocervical distraction.

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In 1830, Scottish physician Charles Bell first described nontraumatic atlantoaxial rotary subluxation in a patient with syphilis and pharyngitis that contiguously traveled to the cervical spine.¹ The patient expired from subsequent extension of the infection to the epidural space, causing spinal cord compression. Not until 1951 did French otorhinolaryngologist P. Grisel coin the syndrome in his publication of two cases with recent pharyngitis.² Grisel syndrome refers to nontraumatic atlantoaxial rotary subluxation after head and neck infections or procedures. Disruption of the ligamentous complex at C1–C2 causes the head and C1 vertebrae to swivel along a relatively stationary C2 and subaxial spine, resulting in a unique rotational subluxation of C1 on C2.

The literature suggests a rare incidence of approximately 1:100,000,000 per year without gender predilection.³ Known as a pediatric disease, an estimated 68% of cases involve children younger than 12 years, and 90% of subjects are younger than 21 years. Children typically respond well to conservative management, with only a

small percentage requiring surgical intervention. Far less common, Grisel syndrome in the adult population has been sparsely published in the literature.⁴ This case lesson describes unique challenges in the surgical correction of advanced-stage Grisel syndrome in an adult (Fig. 1). This case lesson has been reported in line with the Preferred Reporting of CasE Series in Surgery (PROCESS) guideline.⁵

Illustrative Case

Patient History and Examination

A 50-year-old man with past medical history of diabetic neuropathy and cocaine abuse developed a callus of his left great toe secondary to decreased foot sensation. The patient excised the callus himself without medical attention. The toe progressed to osteomyelitis, requiring surgical amputation at an outside hospital. Having neglected his wound for more than a year, the patient eventually presented to an outside hospital with septic shock and was treated

ABBREVIATIONS CT = computed tomography; OAA = occipital-atlantoaxial.

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* M.M. and J.P. contributed equally to this work.

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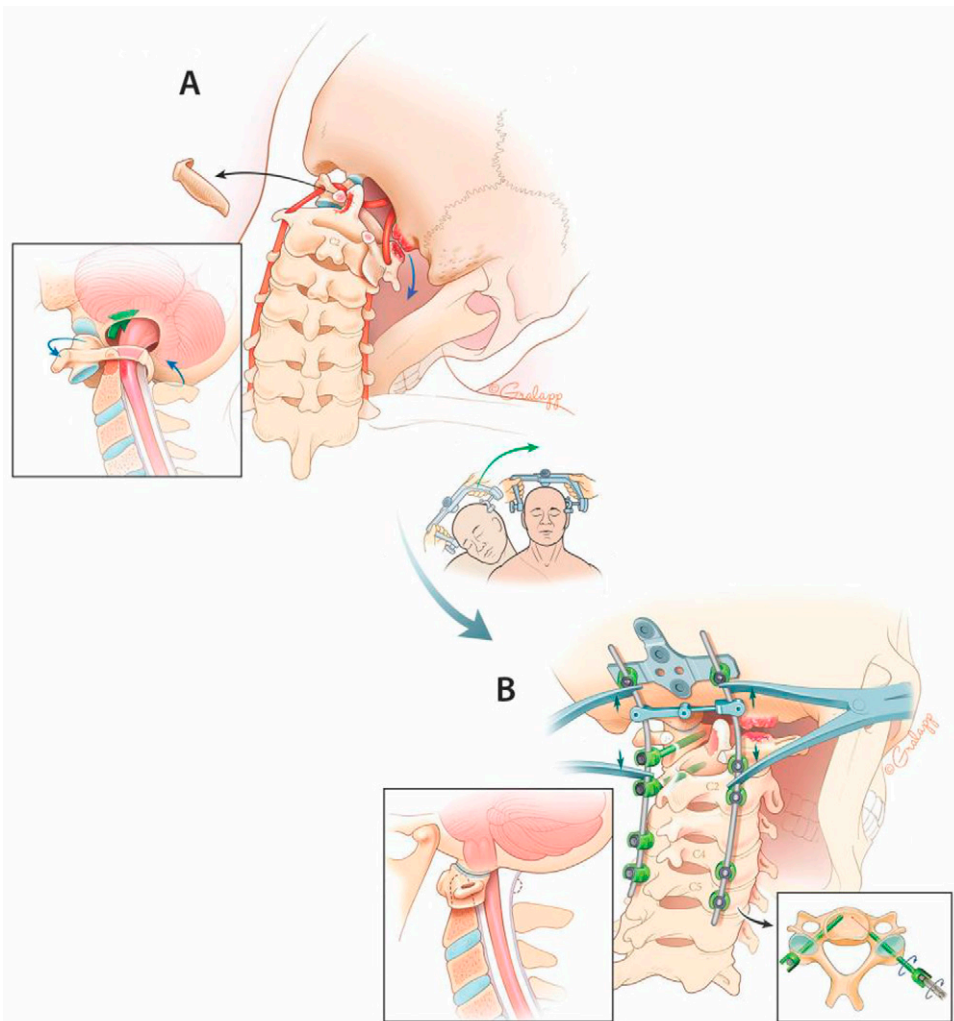


FIG. 1. Illustration of the surgical correction for Grisel syndrome in the adult patient. **A:** Preoperative view: nontraumatic atlantoaxial rotary subluxation with the head rotated toward the right. The inset demonstrates the kinking at the left brainstem–spinal cord junction because of rightward rotation and lateral displacement of the head. **B:** Postoperative fixation: the distractors on the occipital plate and C2 pedicle screws (trajectory of cervical pedicle screws shown in the inset) allow for correction of the cervical deformity. Notice the inset reveals realignment of the atlantoaxial segment and brainstem–spinal cord junction. Illustration by Chris Gralapp.

for methicillin-resistant *Staphylococcus aureus*. A few weeks later, he presented to an outside hospital with a several-month history of severe neck pain from a progressive cervical deformity. Imaging findings prompted transfer to our hospital.

On physical examination, the head was rotated down to the right from severe torticollis. Weakness was noted in the left deltoid (3/5), biceps (4/5), triceps (4/5), and hand grip (4/5). Deep tendon reflexes in the bilateral upper and lower extremities were 3+. He had negative Babinski and Hoffmann's signs. Severe tenderness was elicited on the posterior neck near the craniocervical junction. No obvious fluctuance was appreciated. The left great toe was amputated, and the wound healed by secondary intention. The left foot appeared mottled with poor peripheral pulses.

On admission to our institution, blood culture results remained negative. C-reactive protein was initially 2.9 and erythrocyte

sedimentation rate was 86. White blood cell count was only mildly elevated to 10.7.

Imaging findings were concerning for a soft tissue phlegmon of the neck extending into the cervical spinal column. Computed tomography (CT) demonstrated disarticulation of the atlantoaxial joints bilaterally, causing the calvaria and C1 to rotate to the right along the axial and coronal plane (Fig. 2A–C). The occiput and atlas were completely subluxated to the right of the fixed, in-line C2, which is consistent with rotational atlantoaxial subluxation. The left C1 lateral mass–posterior arch junction eroded into the dens and C2 vertebral body, and the right C1 lateral mass was laterally subluxated relative to C2. Magnetic resonance imaging revealed an infectious process at both the right occiput–C1 joint and bilateral atlantoaxial joints with retrodental epidural extension (Fig. 3A). The rightward displacement of the head had caused significant kinking



FIG. 2. Preoperative CT. Sagittal CT demonstrates (A) destruction of the joint between the right occipital condyle and C1 superior articulation and (B) migration of the left C1 lateral mass anterior to the midline dens. Coronal CT demonstrates (C) the right lateral bending along the coronal plane, which caused the left C1 posterior arch to erode through the dens and C2 vertebral body. **D and E:** C4 vertebrae reveal generous pedicle sizes, well-suited for subaxial pedicle screws.



FIG. 3. Preoperative magnetic resonance imaging (MRI). **A:** Notice epidural extension of the abscess on the preoperative MRI. Axial (B) and sagittal (C) MRI reveals a left-sided contusion at the cervicomedullary junction. Compression occurs because both the head and C1 are rotated and laterally displaced against the dens (at midline position). This rightward displacement kinks and contuses the left junction of the brainstem–spinal cord.

of the lower brainstem–upper cervical spinal cord, with the dens severely compressing and contusing the left side of the cervicomedullary junction (Fig. 3B and C). CT with contrast was concerning for a left vertebral artery stretch injury in which the C1 and C2 transverse foramina were splayed.

Operation

Posterior cervical approach was planned for open reduction and internal fixation to restore the bony alignment and protect the neural elements. Awake fiberoptic intubation was planned. After short-acting succinylcholine was administered, no further paralytics were included in the anesthetic portfolio. Neuromonitoring included somatosensory evoked potentials, motor evoked potentials, and electromyography. After confirming a train-of-four at the ulnar nerve, a Mayfield clamp was placed along the axial plane above the pinna. Using extremely controlled movements, axial in-line traction was carefully introduced in an attempt to realign the skull with the long axis of the neck. We believe that the traditional method of gradual axial traction on the floor over a few days is generally not well tolerated by patients. In addition, this condition tends to yield itself to more immediate reduction because this syndrome is typically less chronic and the neck less rigid.

The patient was turned prone on gel rolls onto the operating table. The head was positioned under fluoroscopy to ensure the hard palate was perpendicular to the floor (Fig. 4A). This position ensures that the occipitocervical fusion fixes the patient's line-of-sight in a plane parallel to the ground. After a sterile prep and drape, the skin was scored from theinion down to the vertebrae

prominens. Monopolar cautery was used to incise the ligamentum nuchae along the midline, avascular plane until the cervical spinous processes were reached. The soft tissues were dissected off the bone in a subperiosteal fashion until the lateral masses were completely exposed. Because the posterior atlantooccipital membrane

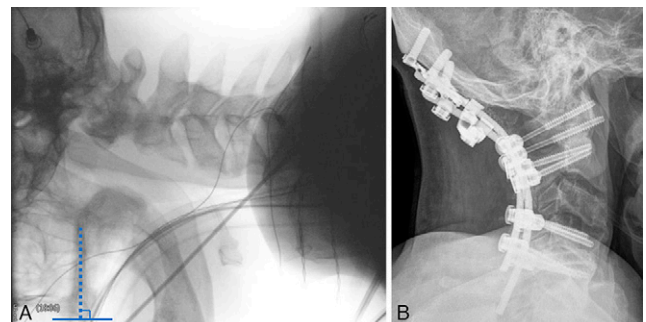


FIG. 4. **A:** Intraoperative fluoroscopy confirms that the hard palate (dotted line) is perpendicular to the floor (straight line), which ensures a neutral gaze in occipitocervical fusions. **B:** Postoperative radiograph illustrating realignment of the head with the cervical spine. The lateral image reveals the surgeon's preference for rod contouring. Also, notice that the superior direction of the C1 lateral mass and C2 pedicle screws shifts to an inferior direction of the C4 and C5 pedicle screws.

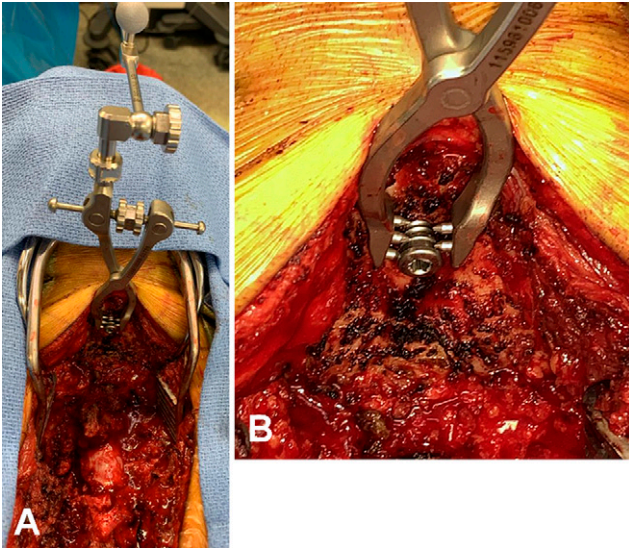


FIG. 5. The proud occipital screw without the occipital plate serves as an anchor for the navigated reference frame.

was disrupted, the tissues were carefully dissected at the level of the foramen magnum. Subperiosteal dissection at C1 has a 1-cm safety zone from midline before the vertebral artery becomes at risk of injury. The occipital bone was widely exposed up to the inion to allow ample surface area for occipital plating, although Figure 1B illustrates that realignment of the occipitocervical junction C1 laminectomy was performed to adequately protect the cervicomedullary junction during bony reduction.

Cranio-cervical fixation was planned with the O-arm (Medtronic, Inc.). First, a temporary screw was inserted at the expected position of the upper occipital screw to anchor the navigated reference frame (Fig. 5). We find this method to be superior in providing more accurate registration and achieving more reliable navigation because it avoids registration errors related to the natural mobility of the cervical segments.

After intraoperative CT acquisition of the craniocervical junction, fixation was begun by insertion of C1 screws at the midplane of the lateral mass mediolaterally. Because of concern for stretch injury of the left vertebral artery (Fig. 1), the C1 screw is typically used because it tolerates up to 20° of medial angulation, allowing the surgeon to confidently avoid injury to the remaining, intact right vertebral artery.⁶ The rostrocaudal trajectory should remain exactly parallel to the posterior C1 arch, approximately 20° in the cephalad direction. Although C1 screws are often omitted in occipitocervical fusions because of difficulty persuading the rods into the deeper and more medial position of the C1 tulip relative to the remaining construct, we typically insist on using them. Bilateral C1 fixation is of utmost importance in rotational atlantoaxial subluxation, as in Grisel syndrome. The three surgical pearls for C1 screw positioning to facilitate connection to the rod are as follows: (1) medialization of the C1 screw to lateralize the tulip, (2) placement of 10-mm or longer C1 screws to place the tulip in a more superficial position to be able to connect the tulip to the occipital rods, and (3) use of extended (reduction) tabs to capture any rod not in line with the screw. An offset joint can be used as a bailout technique.

In our patient, the subaxial spine exhibited generous pedicle anatomy (Fig. 2D). In occipitocervical fusions for rotational atlantoaxial subluxation in Grisel syndrome, the stronger pedicle screws confer several advantages over weaker lateral mass screws.⁷ The higher load-to-failure of the pedicle screw allowed for a more perfect sagittal alignment (later in our case). Because of the long-standing preoperative torticollis, the greater pullout strength of the pedicle screw yields the more rigid construct necessary for any residual neck bending postoperatively.

Adequate lateral exposure is required for safe placement of cervical pedicle screws. C2 pedicle trajectory began just a few millimeters lateral to the midpoint of the pars in the axial plane and in line with the superior edge of the lamina in the sagittal plane.⁸ The screw was directed 30° to 45° medially and superiorly. The trajectory changed in the subaxial spine. Although bilateral hemilaminotomies, if not full laminectomies, are recommended for these cervical pedicle screws, neuronavigation obviated any bony resection necessary to manually visualize the anatomy of the pedicle, and this has become the standard of care for this technique. For the subaxial spine, just below the facet joint in the craniocaudal direction, the starting point of the pedicle screw sat at the mediolateral midpoint of the lateral mass. The trajectory occurred 45° medially as well as perpendicular to the lateral mass in the caudal direction (Fig. 2E). Instrumentation was terminated at C5.

Both the navigation reference frame and the occipital screw were removed. The occipital plate was secured below the level of the inion at the external occipital crest, or medial nuchal line. (Occipital screws are anchored at the occipital keel where the occipital bone is thickest.) A contoured rod was used to connect the occipital bone to the cervical spine. All the cervical set screws were tightened. Final correction of the deformity in the sagittal plane was achieved by maneuvering the occipital end of the rod against the occipital screws (Fig. 4B). The occipital set screws were given a second and final tightening. A cross-link was applied to prevent torsional deformation of the construct, and bone graft in the lateral gutters was placed to augment long-term arthrodesis. After meticulous hemostasis, the muscle, fascia, and skin were reapproximated in a multilayered closure.

Outcome

One week after surgery, the patient was discharged in stable condition. A few months after surgery, the patient's left arm monoparesis had fully resolved, and he had developed bony fusion across the occipital-atlantoaxial (OAA) joints with no signs of hardware failure or recurrence of subluxation or torticollis.

Discussion

Observations

Here we report a case of severe adult Grisel syndrome, characterized by inflammatory rotational subluxation and spondylitis of the OAA joints. According to the Fielding and Hawkins classification system, disruption of bilateral C1-C2 facet joints and the transverse ligament would be described as a type III rotatory subluxation.⁹ There have been only a handful of previously reported cases of severe Grisel syndrome in adults, all of which required surgical stabilization.^{10–13} Grisel syndrome more commonly presents in children or adolescents and often results from oropharyngeal infections or is preceded by ear, nose, and throat procedures.¹⁴ In children, the subluxation is commonly due to ligamentous laxity without frank

osseous destruction of the OAA articulations. The rarer condition in adults has also been called atlantoaxial subluxation of inflammatory origin or osteomyelitis of the atlantoaxial joints with rotatory subluxation. Some authors draw a distinction between the milder pediatric Grisel syndrome and the more severe syndromes of destructive OAA osteomyelitis with atlantoaxial subluxation and torticollis observed in adults.¹⁰ For the ease of discussion, we refer to both entities simply as pediatric and adult Grisel syndrome. Our case was notable for the severity of the pathology, the unusual presentation in an adult patient with no prior oropharyngeal infections or procedures, and the specific operative techniques required for final stabilization.

Management of pediatric Grisel syndrome is primarily observational, with treatment focusing on immobilization in a soft collar and antibiotic therapy. A systematic review of 171 cases of pediatric Grisel syndrome found that 96% of children were initially managed conservatively, with only 12% requiring surgery as either a first- or second-line treatment.¹⁵ Comparatively, adult Grisel syndrome, although much rarer, is more frequently managed surgically. Kerolus et al. reported two cases of C1-C2 instability that ultimately required surgical fixation. These cases were managed with C1-C2 fusion and C1-C4 fusion, respectively. Their literature review of Grisel syndrome in adult patients identified 14 cases, and of those, only two patients were successfully treated conservatively.¹² These authors suggest that in adult patients with atlantoaxial instability, early surgical management is warranted. Yamazaki et al. reported a case of adult Grisel syndrome in which the patient was treated with occipitocervical fusion after failing 3 months of conservative therapy with halo vest.¹⁶

Yamane et al. described a similar case of adult Grisel syndrome with right-sided torticollis and quadriparesis that was effectively treated with occipital-C4 fusion.¹¹ Occipitocervical fixation is the treatment of choice in cases of severe Grisel syndrome with complete dislocation of the atlantoaxial joints as well as involvement of the atlantooccipital joints.^{11,13} Ishikawa et al. reported a case of adult Grisel syndrome with Fielding class IV atlantoaxial instability requiring antibiotics, halo fixation, and ultimately, occiput-C3 fixation.¹³ Similar to our case, the diagnosis of Grisel syndrome was delayed by 2 months after the onset of neck pain, and the pathology had spread to involve the atlantooccipital joint as well. Halla et al. reported on a similar case of adult Grisel syndrome with delayed diagnosis, painful leftward rotational torticollis, and acute right-sided hemiparesis.¹⁰ This case is similar to that of our patient, who developed right-sided painful torticollis and left arm weakness. These reports suggest that in patients who present with a painful, fixed torticollis and contralateral neurological deficits, atlantoaxial subluxation should be suspected.

The dichotomous presentation and management of pediatric and adult Grisel syndrome is largely due to important distinctions in the pathophysiology of each disorder. Pediatric Grisel syndrome has been suggested to be caused by a two-hit hypothesis, in which children with preexisting laxity in the atlantoaxial ligamentous complex develop muscle spasms and a painful torticollis in the setting of a cervical inflammatory process.¹⁷ The muscle spasm, in response to inflammation, creates a rotational subluxation in a child with preexisting ligamentous laxity. These patients rarely have spondylitic or destructive changes in their atlantoaxial joints and therefore commonly respond to conservative management. This condition is often preceded by oropharyngeal infections or procedures because the pharyngovertebral veins act as a conduit for bacteria or inflammatory mediators to travel between the pharyngeal space and periodontal

venous plexus.¹⁸ Comparatively, adult Grisel syndrome is often characterized by frank osteomyelitis of the OAA articulations, weakening of the ligamentous constructs, and gross instability that results in a rotatory subluxation of the occipitocervical junction. The risk factors for adult Grisel syndrome, such as a history of diabetes or systemic bacteremia, are similar to those for osteomyelitis in other parts of the body. By comparison, adult Grisel syndrome is more likely to require surgical stabilization because it implies an inherently more severe pathology.

Regarding management strategy, the approach should be dictated by the extent of the pathology. In mild cases of adult Grisel syndrome with Fielding class I subluxation only and no torticollis, hard collar or halo immobilization may be sufficient. More severe cases should be considered for early surgical fixation. When the inflammatory process is isolated to the atlantoaxial joints only, C1-C2 fusion may be sufficient. In patients with significant subluxation but mild or reducible torticollis, standard occipitocervical fusion is likely adequate to provide stability and bone fusion. In patients who present with severe, fixed torticollis and involvement of multiple OAA joints, stronger fixation should be considered, including subaxial pedicle screws. Subaxial pedicle screws are superior to lateral mass screws in terms of pullout strength and likelihood of fusion.¹⁹ The stronger three-column fixation can aid in correction of cervical deformity and decrease the likelihood of postoperative hardware failure.

Lessons

In conclusion, nontraumatic atlantoaxial rotary subluxation, as in the infectious process in Grisel syndrome, is uncommon in adults. Even rarer are adult patients who require operative intervention for Grisel syndrome. This case describes the unique surgical pearls necessary for the occipitocervical fusion of an unstable craniocervical junction with compression at the brainstem–spinal cord junction.

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Author Contributions

Conception and design: Macki. Acquisition of data: Macki, Pawloski. Analysis and interpretation of data: Macki, Pawloski, Abdulhak. Drafting the article: Macki, Pawloski, Fadel. Critically revising the article: Macki, Pawloski, Fadel. Reviewed submitted version of manuscript: all authors. Approved the final version of the manuscript on behalf of all authors: Macki. Statistical analysis: Macki. Administrative/technical/material support: Macki. Study supervision: Macki, Abdulhak.

Correspondence

Mohamed Macki: Henry Ford Hospital, Detroit, MI. mmacki2@hfhs.org.